Primary Huge Exophytic leiomyosarcoma of the stomach: A care report and review of literature

Abstract

Gastric leiomyosarcoma accounts for 1% of all gastric tumors. This type of tumor typically arises from the muscularis propria and is most often found in the body, fundus, cardia, and pyloric antrum of the stomach. Gastric leiomyosarcoma commonly presents as a polypoidal mass, exophytic mass, or ulcerative mass. The primary symptoms include bleeding leading to hematemesis, anaemia, abdominal pain, weight loss, and the presence of an abdominal mass.

Effective diagnostic modalities include endoscopic ultrasonography, contrast-enhanced CT scan, and MRI. Immunohistochemically, the tumoral cells in gastric leiomyosarcoma show positive immunoreactivity to smooth muscle actin and desmin while testing negative for CD117 (C-kit) and CD34. The most effective treatment is complete surgical resection with negative margins, typically achieved through either open or laparoscopic partial gastrectomy.

We report the case of a 57-year-old male who presented with a large mass in the epigastric and left hypochondriac region. Diagnosis was confirmed via CT scan of the abdomen, and histopathological examination verified a high-grade gastric leiomyosarcoma.

Key words

Leiomyosarcoma, stomach tumor, gastric tumors, GISTs.

Introduction

Leiomyosarcoma is a tumor originating from smooth muscle and is an important differential diagnosis from gastrointestinal stromal tumors (GISTs). In the late 1990s, definitive immunohistochemical methods were developed to distinguish GISTs from leiomyosarcomas. The diagnosis of primary leiomyosarcoma now relies on a combination of histopathological features, positive immunoreactivity to smooth muscle antigens, and non-responsiveness to GIST immune markers, specifically CD117 (C-kit), CD34, and DOG1. Currently, primary leiomyosarcomas of the gastrointestinal tract, including gastric leiomyosarcomas, are rare. [1,2,3,4]

The etiology of gastric leiomyosarcoma has been associated with factors such as ionizing radiation, Epstein-Barr virus, and various chemical exposures. There is also a potential role of Helicobacter pylori in its development. [2,4,5]

Accurate diagnosis relies on histological examination with immunohistochemical analysis. This aggressive disease does not respond to chemotherapy or radiation; surgery is the only curative treatment. Leiomyosarcoma typically spreads hematogenous to the liver and lungs. The overall 5-year survival rate ranges from 16% to 56%, and recurrence occurs in 36% to 60% of cases, typically within two years after resection. Surgical resection remains the treatment of choice for gastric leiomyosarcoma. [1,5,6,7]

Case Report

A 57-year-old male was admitted to our center on January 15, 2020, presenting with complaints of an abdominal lump and pain in abdomen for the past six months. Physical examination revealed normal vital signs. There was no history of hematemesis, jaundice, or vomiting. Abdominal examination identified a large palpable mass in the epigastrium and left hypochondrium, with mild tenderness. Blood tests, including liver and renal function tests, were within normal limits.

An abdominal CT scan revealed a solid, exophytic, nodular mass measuring 25 x 15 x 10 cm. The mass was ill-defined, heterogeneous, and lobulated, with its main bulk in the left hypochondrium, originating from the gastric body. It exerted pressure on the stomach, spleen, and pancreas and diagnosis was gastric leiomyosarcoma.

We performed an exploratory laparotomy via a midline abdominal incision with the aim of complete tumor removal, which is the standard treatment for gastric leiomyosarcoma. The patient underwent a partial gastrectomy, removing the large 25 x 15 x 10 cm multinodular mass with a 5 cm margin of normal gastric tissue along with the greater omentum.

Gross examination revealed a multinodular mass weighing 3.5 kg. Microscopic examination showed spindle-shaped tumor cells with hyperchromatic pleomorphic nuclei and a high mitotic index. Immunohistochemistry was positive for smooth muscle actin and desmin, but negative for CD117.

The patient was discharged on the tenth post-operative day and was advised to pursue chemotherapy at a cancer hospital due to the high-grade nature of the gastric leiomyosarcoma. Unfortunately, the patient passed away one year after surgery at home. **(Fig 1-10)**

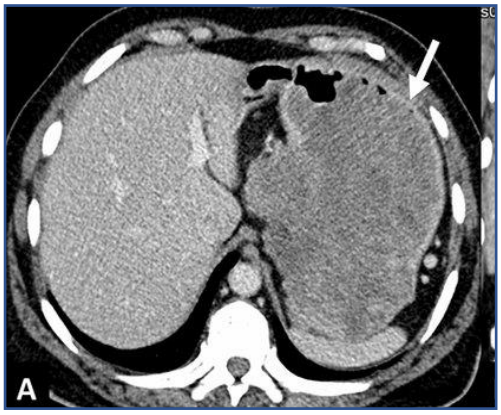

Fig-1 CT abdomen showing a large heterogeneously enhancing exophytic mass arising from the body of the stomach

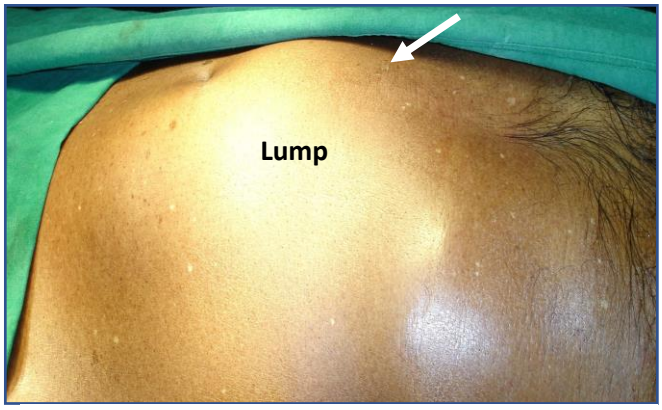

Fig-2 Photograph showing a huge lump at epigastrium and Left hypochondrium

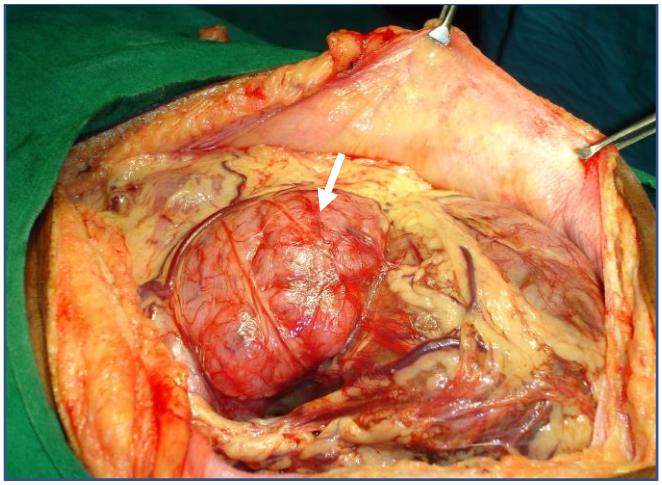

Fig-3 Intraoperative photograph showing an exophytic mass at epigastrium

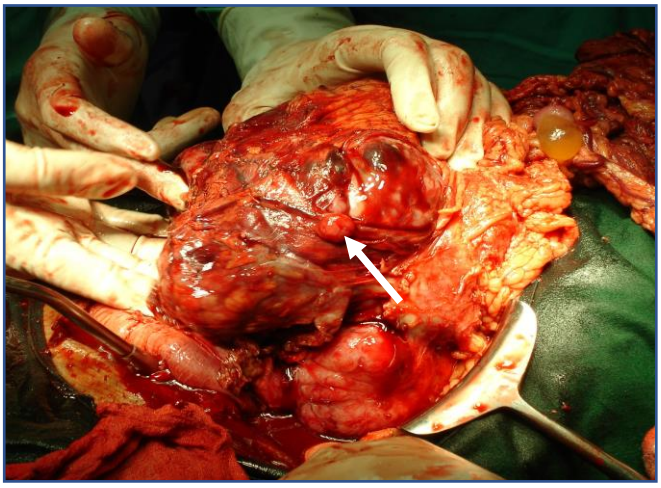

Fig-4 Intraoperative photograph showing a huge, multinodular mass

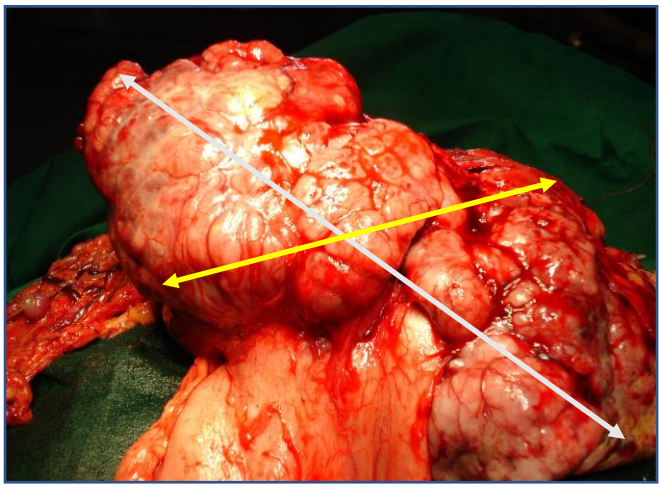

Fig-5 Intraoperative photograph showing a huge multinodular mass arising from body of the stomach

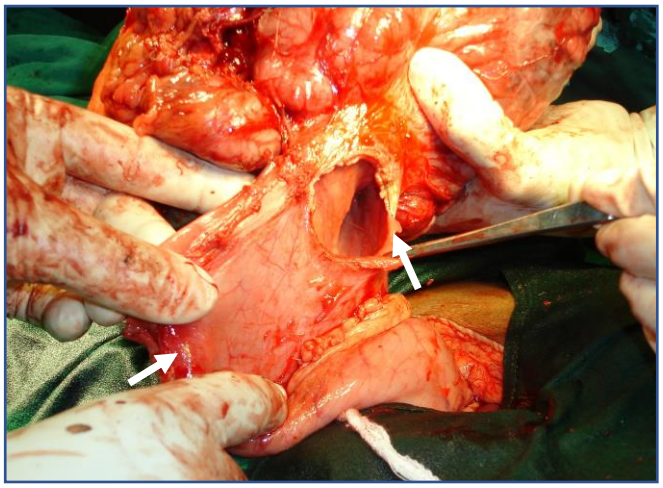

Fig-6 Intraoperative photograph showing a wide excision of gastric mass

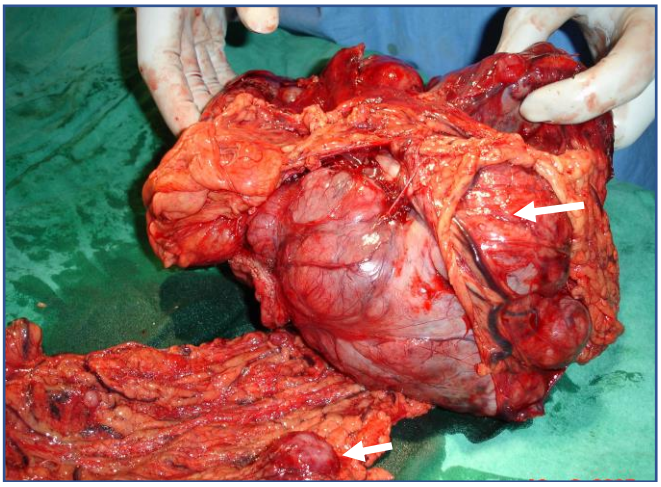

Fig-7 Intraoperative photograph showing a large, polypoidal mass with omental excision

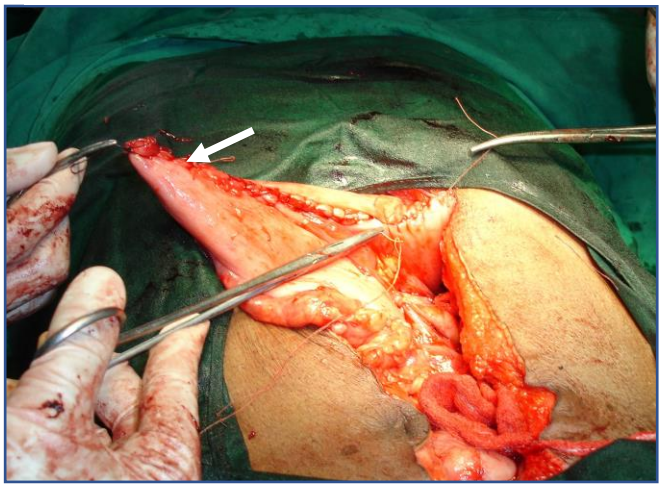

Fig-8 Intraoperative photograph showing a partial gastrectomy done

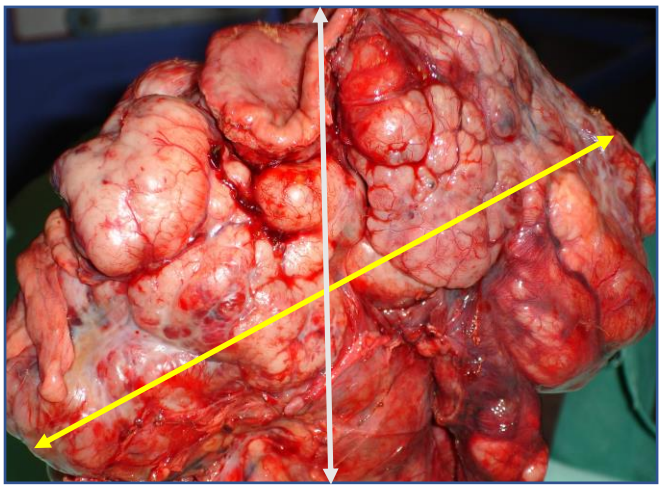

Fig-9 Photograph showing a large multinodular mass measuring 25x15x10 cm weighing 3.5 kg

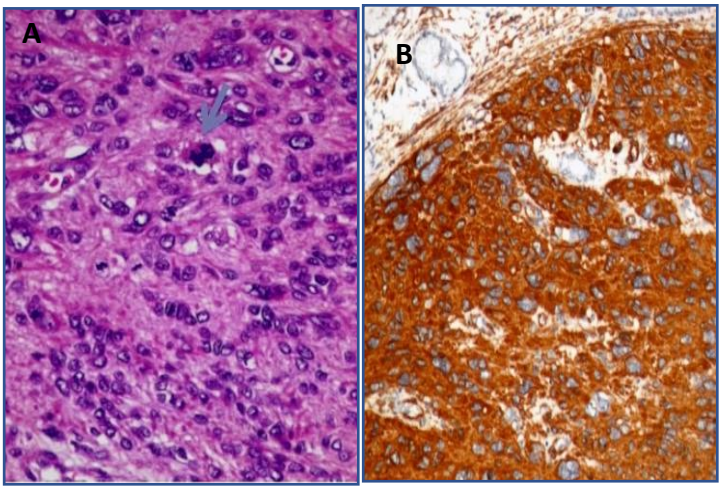

Fig-10 A. Photograph showing smooth muscle cells with nuclear hyperchromasia and numerous mitoses.
B. Well differentiated leiomyosarcoma, neoplastic cells positive (brown) for desmin

Discussion and review of literature

Gastric leiomyosarcoma typically originates from the muscularis propria or muscularis mucosa layer in various parts of the stomach, including the body, fundus, antrum, cardia, and pylorus. Common clinical signs and symptoms include abdominal pain, melena, weight loss, and the presence of an abdominal mass. Tumor sizes can range from 1 to 18 cm. [1,2]

Effective diagnostic tools include endoscopic ultrasonography, contrast-enhanced CT scans, and MRI. These imaging modalities help in detecting and assessing gastric leiomyosarcomas. For definitive diagnosis, histopathological examination is essential. Simple endoscopic biopsies might be inadequate; therefore, endoscopic ultrasound-guided fine needle aspiration cytology is recommended for accurate diagnosis, with a success rate of up to 97%. [3,4,7]

On contrast-enhanced CT, 50% of gastric leiomyosarcomas are intramural, 25% are exophytic, and 15% are endogastric. Large exophytic lesions can cause palpable lumps and exert extrinsic mass effects on adjacent organs. Exophytic leiomyosarcomas are typically large, lobulated masses. Hematogenous spread to the liver and lungs occurs in about 40% of cases, while regional nodal metastasis is uncommon. [5,6,8]

Since the early 2000s, only 19 cases of gastric leiomyosarcoma have been reported, with our case being the 20th. The condition shows a 2:1 male predominance and typically affects individuals between 50 and 70 years of age. Approximately 90% of these tumors arise from the body or fundus of the stomach, with 10% originating from the gastric antrum. [2,7,10]

Immunohistochemistry is crucial for differentiating between GISTs and leiomyosarcoma. Leiomyosarcomas are identified by their positivity for smooth muscle actin or desmin and negativity for CD117, DOG1, and CD34. [1,2,4]

Kawaguchi et al. found that a combination of gemcitabine and docetaxel could induce regression in gastric leiomyosarcoma, suggesting a potential therapeutic option. In advanced cases, chemotherapy with agents like doxorubicin and ifosfamide is used. Second-line therapies include trabectedin and pazopanib, which inhibit multiple tyrosine kinases. [2,3]

Immunotherapy with agents such as nivolumab and pembrolizumab is also being explored. Ongoing phase I and II trials are investigating new drug combinations with radiation and chemotherapy. [2,6]

Surveillance imaging with CT or MRI of the chest, abdomen, and pelvis is recommended every six months for the first two years and then annually thereafter. The main prognostic factors include histopathological grading, tumor size (>5-7 cm), evidence of synchronous metastasis, and gastric wall infiltration. The 5-year survival rate for patients with gastric leiomyosarcoma is approximately 22%. [1,2,4]

Treatment

Surgery is the only curative option for gastric leiomyosarcoma. The type of surgery depends on the tumors size and location and may range from wedge resection to partial or total gastrectomy, with En-bloc resection of adjacent organs if involved. Complete surgical resection with negative margins is considered the most effective approach. [1,2,3]

Methods such as laparoscopic gastrectomy and endoscopic submucosal dissection are used for tumor removal. In March 2018, Stato et al. reported the first case of small gastric submucosal leiomyosarcoma treated with endoscopic submucosal dissection. A combination of laparoscopic

resection and endoscopic determination of cutting lines and tumor margins has shown efficacy. In 2021, Toru Takagi et al. documented a successful combination of laparoscopic and endoscopic approaches to treating intraluminal gastric leiomyosarcoma. [4,6,8]

In cases where the tumor has metastasized, treatment options may include chemotherapy, chemoembolization, and high-intensity focused ultrasound therapy. Multimodal therapy involving radiotherapy (RT) and chemotherapy is being inverted and currently recommended for cases with high local recurrence and high grade, high rick leiomyosarcoma. In patient with metastatic disease, surgical resection in often recommendation for palliative purpose. [1,2,5,6]

Conclusion

Gastric leiomyosarcoma are extremely rare and diagnosis is based on histological examination with immunohistochemistry for marking and accurate diagnosis. The standard treatment for gastric leiomyosarcoma is complete resection of the tumor, a wedge resection, partial or total gastrectomy with En-bloc resection if adjacent organs are involved.

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